Friedreich's Ataxia (FA)

Friedreich's ataxia is a genetic, progressive, multisystem disease that causes ataxia, neuropathy, spasticity, cardiomyopathy, cardiac arrhythmias, scoliosis, fatigue, diabetes, vision and hearing loss, and other symptoms. Severity and progression of symptoms vary between individuals. For most, progressive ataxia results in continued loss of mobility and independence.

Individuals with FA may present to the ER for several reasons including chest pain, cardiac arrhythmia, hypotension, infection (such as viral or urinary tract infection), a recent fall, pain, or a diabetic emergency

Cardiac Disease

- Abnormal EKGs (negative T wave) and elevated troponin levels are common in people with FA at baseline and do not typically indicate the presence of acute coronary syndrome.
  - Ask to see a copy of patient's most recent EKG and troponin levels to determine baseline.
  - Coronary artery disease is not more common in people with FA.
- Cardiomyopathy is common and can progress to heart failure.
- Arrhythmias seen in FA include SVT, atrial fibrillation, and atrial flutter.
  - These arrhythmias should be addressed with standard treatment.

Fluids, Surgery and Anesthesia

- In an emergency setting, fluids should not be avoided if indicated to support blood pressure, however, fluid balance should be carefully monitored.
- Individuals with FA are at a greater risk of cardiovascular instability and complications during major surgery.
  - Careful monitoring of heart function and fluids is necessary.
  - The heart may not tolerate fluid shifts during the surgical procedure, long periods of anesthesia, or low blood pressure.
  - A multidisciplinary approach to surgery is recommended.
- A wide range of anesthetic options can be used for individuals with FA, including spinal nerve blocks or anesthetics given intravenously.

Emergency Information for Individuals with Friedreich's Ataxia

Patient name:_____

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Emergency contact:_____

_____

Phone number:_____

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Cardiologist:_____

_____

Phone number:_____

_____

Neurologist:_____

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Phone number:_____

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Primary care physician:_____

_____

Phone number: _____

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Important Medical Information
(current medications and doses, medical diagnoses, etc):

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This pamphlet summarizes information from the FA Clinical Management Guidelines. To access the full guidelines, scan the QR code

or visit frdaguidelines.org

FARA Friedreich's Ataxia Research Alliance